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Fuchs' spot in degenerative myopia: Clinico-pathological aspects and laser treatments

Abstract

This article reviews representative literature on the epidemiology, clinical observation, pathological examination, and laser treatment of Fuchs' spot in degenerative myopia. Degenerative myopia is the seventh leading cause of registered blindness in adults in the United States. Fuchs' spot is one of the major complications of degenerative myopia. With correct evaluation of the degenerative change in the macula of the myopic eye, appropriate vision care can be provided to reduce the probability of blindness. Fluorescein angiography is of great value in the detection and localization of subpigment neovascular membranes. Laser photocoagulation shows a beneficial effect on the final visual result in some cases. Fuchs' spot results from two different stages in the development of a hemorrhagic disciform macular detachment in myopia. A knowledge of the exact mechanism of chorioretinal degenerations in the myopic eye must await future research.

Degree Type

Thesis

Degree Name

Master of Science in Vision Science

Committee Chair

Salisa Williams

Subject Categories

Optometry

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FUCHS' SPOT IN DEGENERATIVE MYOPIA :
CLINICO-PATHOLOGICAL ASPECTS AND LASER TREATMENT

By
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A thesis submitted to the faculty of the
College of Optometry
Pacific University
Forest Grove, Oregon
for the degree of
Doctor of Optometry
January, 1993

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ACKNOWLEDGEMENTS

I would like to thank Dr. Salisa Williams for her knowledge and time on my thesis. I would also like to thank Pacific University College of Optometry for the use of their facilities.

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ABSTRACT

This article reviews representative literature on the epidemiology, clinical observation, pathological examination, and laser treatment of Fuchs' spot in degenerative myopia. Degenerative myopia is the seventh leading cause of registered blindness in adults in the United States. Fuchs' spot is one of the major complications of degenerative myopia. With correct evaluation of the degenerative change in the macula of the myopic eye, appropriate vision care can be provided to reduce the probability of blindness. Fluorescein angiography is of great value in the detection and localization of subpigment neovascular membranes. Laser photocoagulation shows a beneficial effect on the final visual result in some cases. Fuchs' spot results from two different stages in the development of a hemorrhagic disciform macular detachment in myopia. A knowledge of the exact mechanism of chorioretinal degenerations in the myopic eye must await future research.

INTRODUCTION

Degenerative or pathologic myopia is often defined as that state in which a number of serious complications are associated with an excessive axial elongation of the eye. A wide spectrum of fundus changes can be seen in these cases. Fuchs' spot is one of the major complications of degenerative myopia. This pigmented lesion in the macula of the myopic eye was first described by Forster in 1864, and later studied extensively by Fuchs in 1901. Since the original description, common usage considers Fuchs' spot as any dark spot at the posterior pole of the patient with high myopia.

Fuchs' spot is not uncommon in the myopic population. It is generally believed to affect 5% to 10% of patients with myopia exceeding 5 diopters (Campos, 1957; Curtin, 1971). The lesion commonly occurs during the productive years of young adulthood. It initially manifests itself as a disciform response, and later becomes a flat dark spot in the macula. Central vision is often severely reduced in these cases.

A survey conducted by the National Society for the Prevention of Blindness (1966) showed that degenerative myopia is the seventh leading cause of registered blindness in adults in the United States. It has been estimated that the frequency of degenerative myopia is about 1% in the general population (Duke-Elder, p241; Curtin, p228; Ryan, p201; Sperduto et al, 1983; Hu, 1987). The socioeconomic impact of myopic blindness is markedly greater when this statistic is considered (Perkins, 1979).

It is the purpose of this paper to review available literature pertaining to Fuchs' spot - one of the chorioretinal degenerations occurring at the posterior pole of the myopic eye, and to suggest that with the correct evaluation of the condition, appropriate vision care can be provided to reduce the probability of blindness. Areas to be discussed include clinico-pathological aspects and laser treatment of the lesion.

ANATOMY AND HISTOLOGY OF THE MACULAR REGION

The macula lutea is a shallow oval depression with a diameter of about 1.5 mm in the center of the posterior retina. It lies 3.5 mm temporal to the edge of the disc and about 1 mm inferior to the center of the disc (Warwick, p137) . The yellow coloration of the macula is due to the presence of a lipid-soluble pigment, xanthophyll, which is present in greatest concentration in the outer plexiform layer (Fine, p118) . Ophthalmoscopically, the macula often appears slightly darker in color than the remaining fundus. The temporal pairs of retinal vessels arch radially and converge toward the macula but there is a lack of blood vessels in the central area. The boundary of the macula can be noted by a partial or complete annular ring. A tiny bright light reflex may also be detected in the center of the macula (Fine, p113; Snell, p196).

The central retina (area centralis), or macular region, is defined histologically as that area of the posterior retina having more than one layer of ganglion cells. It is about 5.5 mm in diameter, and subdivided into three zones: fovea, parafovea and perifovea (Hogan, p492). The fovea covers an area of 1.5 mm. The sides of foveal depression are called the elivus and the floor of the depression is known as the central fovea or foveola (0.35 mm in diameter). In the central fovea, there are no rod, bipolar and ganglion cells. At the peripheral edge of the fovea, the ganglion cell layer reaches six to eight layers in thickness (Apple, p2). The parafovea, 0.5 mm in width, immediatly surrounds the fovea. It is characterized by the largest accumulation of bipolar and ganglion cells in the entire retina. The perifovea is the peripheral zone of the macular region, and measures 1.5 mm in width. In this area, the density of cones decreases markedly, and the outer plexiform layer changes from Henle's fiber layer to a more usual arrangement.

There are several ways histologically to identify the macular region and its substructures (Apple & Rabb, p15; Fine, p111; Hogan, p491; Warwick, p137) :

1. The central fovea is the thinnest portion of the retina. Its thickness consists mainly of photoreceptors and their nuclei. The

rods here are wholly absent, and the cones are more elongated and slender than those of the extramacular region.

2. In contrast to the central fovea, the border of the fovea is the thickest area of the retina, where the ganglion cell layer is stratified into six to eight layers as opposed to the single layer of nuclei seen elsewhere in the peripheral retina.

3. The outer plexiform layer in the macular region has an unique appearance. The nerve fibers in this layer course obliquely, which is termed Henle's fiber.

4. The blood vessels in the area are mostly capillaries. They are present in the inner retinal layers up to the fovea. There is a capillary-free zone of 0.4 mm in the center of the macula.

5. The pigment epithelial cells are increased and more richly pigmented in the macular region.

The retinal pigment epithelium (RPE) consists of a single layer of cells that extends from the edge of the optic disc to the ora serrata. Typical features of RPE cells are their polygonal shape, tight junctions, apical microvilli, and basal membrane infoldings. Flat sections show these cells to be hexagonal, especially in the central retina. RPE cells are taller and narrower in the macular region than elsewhere (Warwick, p106) . These cells usually contain a rather large nucleus, a well-developed endoplasmic reticulum, a Golgi apparatus, and numerous pigment granules. The basal ends of the cells are markedly folded and rest on the basement membrane of the choroid. The apical parts of the cells show many microvilli measuring about 5 to 7 μm long (Warwick, p105) . These microvilli surround the outer one-third processes of the rods and cones, and there are no specialized attachments between these two structures. The lateral aspect of each cell is united to its neighbors by a large number of zonula adherens and occludens. These tight junctions are essential in maintaining the isolation of the rod and cone receptors from the choroidal circulation.

Bruch's membrane is a thin complex membrane that occupies the plane between the retinal pigment epithelium and the choroidal capillaries. It used to be recognized as a fine amorphous collagenous zone by light microscopy. Electron microscopic study (Nakaizumi,

1964) revealed that Bruch's membrane consists of five different components: (1) the basement membrane of RPE; (2) inner collagenous zone; (3) elastic fiber area; (4) outer collagenous zone; and (5) the basement membrane of the choriocapillaris. The membrane thickens with advancing age. It is thickest in the posterior polar region, and gradually thins anteriorly. In adults, Bruch's membrane measures 2 to 4 μm thick centrally, but only 1 to 2 μm thick in the periphery (Hogan, p332).

The blood supply of the retina is from two sources: the central artery of the retina and the capillaries of the choroid. The central retinal artery is a medium-sized vessel, which exits the ophthalmic artery near the optic foramen and enters the optic nerve about 12 mm posterior to the eyeball (Snell, p171) . It usually divides into four main branches in the optic nerve head: the upper and lower nasal and temporal. These arterial branches course in the nerve fiber layer close to the internal limiting membrane. The capillaries derived from the terminal arterioles form superficial and deep networks, which may extend as deeply as the inner nuclear layer. The capillary networks are well developed in the macular region. However, the central fovea is totally devoid of vessels. The central vein of the retina is formed by tributaries that accompany the corresponding arterioles. The diameter of the artery is about two-thirds to three-quarters that of the corresponding vein.

The capillary layer of the choroid is located next to the retinal pigment epithelium. Its afferent blood supply is mainly from the short posterior ciliary arteries, which enter the choroid around the optic disc and branch extensively as they pass forward and internally into the choroid (Hogan, p363) . The choroidal capillaries differ from those in other organs by their greater diameter of the lumina, which may allow at least two to three red blood cells to pass through at a time. The capillaries in the macular region have very thin walls and multiple fenestrations. The number of vessels here is much greater than elsewhere. The capillaries in the inner layer of the choroid form a distinct lobular pattern. Each lobule is supplied centrally by a precapillary arteriole and demarcated peripherally by

a ring of postcapillary venules (Hayreh, 1973,1975) . The vena vorticosae drain the blood from all parts of the choroid.

ETIOLOGY AND PATHOGENESIS

ETIOLOGY

The genesis of posterior fundus changes in the myopic eye has been debated for many years. Although these lesions are thought to be either due to biomechanical abnormalities or an inherited dystrophic process (Nobel and Carr, 1982; Ryan, V.2, p201, 1989), the answer remains uncertain. The borderline cases still can be encountered in our daily work. An understanding of the exact mechanism of chorioretinal degenerations in the myopic eye must await future research.

Pathologic myopia typically involves a progressive elongation of the eyeball in the anteroposterior axis. This lengthening of the globe usually develops earlier than degenerative changes in the posterior pole. It is this fact that has stimulated continuing interest in exploring the relationship between this elongation and the fundus changes. The myopic eye enlarges in all its dimensions but principally the anteroposterior. Donders (1864) measured the size of five myopic globes, and found that they unexceptionally took an ellipsoidal shape. Choroidal atrophy in the myopic eye was viewed by the pioneer investigator as a consequence of ocular content stretching within the posterior staphyloma. It has been assumed that the same amount of the retina and choroid has to cover a much greater area when the eyeball elongates. The ocular alterations initiated by the mechanical tissue strain are evidenced by a number of distinctive ophthalmoscopic signs :

1. One common manifestation in this regard is the presence of an optic nerve crescent. The crescent is commonly located on the temporal side of the optic disc. Previous histological studies have revealed a disparity in the area of the crescent between the scleral shell and the lamina vitrea complex (retinal pigment epithelium-Bruch's membrane-choriocapillaris) (Duke-Elder, p 317, 1970). In one study, Curtin and Karlin (1971) demonstrated that the incidence of crescent formation is closely associated with increased axial length. Given the fact that the crescent may also be seen in

emmetropic and hyperopic eyes, the optic nerve crescent as an abiotrophic entity was not considered, but, instead, the result of a mechanical stretching. The classical view that the lamina vitrea complex is drawn back into the ectasia at the posterior pole of the eye with the presence of a myopic crescent is supported by their work;

2. The nasal supertraction of the retina, oblique entrance of the optic nerve and Weiss's reflex streak are more characteristic changes of highly myopic eyes (Duke-Elder, p326). In this case, the nasal retina extends beyond the surface of the disc and blurs its nasal border. The combination of this phenomenon along with the temporal crescent, causes the entire optic nerve head to appear as an oval disc tilting in a temporal direction. A bright crescentic retinal reflex may be found on the nasal side of the disc. All of these fundus changes are believed to be due to a displacement of retinal and choroidal tissues in the local area;

3. A relative pallor of the fundus oculi can be seen in high myopia. It may be found in association with an increased visibility of the choroidal vasculature, which is termed fundus tessellation. These fundus changes are thought to be the result of retinal pigment epithelium and choriocapillary thinning within the ectasia of the globe (Duke-Elder, p327; Curtin, 1977; Rabb, 1987);

4. A straightening of the temporal retinal vessels may be found in some cases. It corresponds directly to the stretching of retinal tissue (Rabb, 1987).

The occurrence of posterior staphyloma is characteristic in patients with pathologic myopia. This ocular sign can be best appreciated by using binocular indirect ophthalmoscopy. The staphyloma has a relative sharp margin in the majority of cases, but occasionally, the edge appears as a rounded ridge over which the retinal vessels are bent (Duke-Elder, p329). Certain staphylomas have an unusual appearance, with several steps or terraces found on the nasal side (Curtin, 1977). Scleral thinning within a posterior staphyloma is obvious (Duke-Elder, p310; Curtin, p251). Although the pathophysiology of myopic staphyloma is unknown, it has been suggested that the sclera of myopic eyes is abnormal, has a low

mechanical resistance, and gradually stretches in response to stress such as intraocular pressure (Balacco-Gabrieli, 1982; Curtin, 1982). The ocular rigidity in high myopia has been found to be reduced (Curtin, p282). Scleral indentations matching the course of the large choroidal veins may be observed in some instances (Samuels and Fuchs, 1952).

Progressive distension of the staphyloma allows its choroidal and retinal contents to undergo a greater biomechanical stress in comparison with others. The choroidal vascular tree in the particular area may be consequently compromised by a retardation and diminution of choroidal blood flow. Avetisov and Savitskaya (1977) confirmed by fluorescein angiography that choroidal and retinal circulations are slowed in severe myopia. The ocular pulse originating from choroidal blood flow is found to be considerably depressed in these cases (To'Mey et al, 1981). Hayreh and Baines' studies in monkeys (1973) showed that the macular region in the eye is situated not only on many boundary zones of the short posterior ciliary arteries, but also on all the boundary zones of the vortex veins. Based on the previous pathophysiologic studies in cerebral circulation, Hayreh (1975) has postulated that the macular area is more vulnerable to vascular disorders than other parts of the choroid due to its preferential loss of choroidal perfusion. This concept can be explained by comparing this process to the failure of a farm irrigation system in which the fields at the end of the line are affected most when the main pipe supplies too little water. The central fovea is totally supplied by the choriocapillaris and functionally active. A small reduction of oxygen supply in the macula slows the local metabolism enough so that a variety of chorioretinal degenerations may eventually occur in the focal area (Green and Rabb, 1981).

Degenerative changes in myopic eyes are found to occur frequently within the posterior staphyloma or near its margin (Curtin, 1977). Chorioretinal atrophy often occurs at the edge of the crescent, where considerable biomechanical stress could be expected (Curtin and Karlin, 1971). The atrophic area in myopia can be discrete. However, these lesions have the tendency to merge together

and exhibit a sharp border. A few investigators (Shapiro and Chandra, 1985; Klein and Green, 1988) have reported that the lacquer cracks observed in some cases appeared shortly after resolution of the subretinal hemorrhages and were more extensive than the preceding hemorrhages. Evidence of choroidal neovascularization was not found at the sites where hemorrhages and lacquer cracks occurred and it was concluded that mechanical stretching is a causative factor of lacquer crack lesions, which directly leads to the rupture of the lamina vitrea complex in myopic eyes. The essential feature of the biomechanical concept is that chorioretinal degenerations in myopic eyes are secondary to the distension of the scleral coat of the eye, which is determined largely by increased intraocular pressure.

In contrast to the biomechanical philosophy, the heredodegenerative theory emphasizes the influence of heredity on the occurrence of myopic fundus changes. A few general observations include :

1. There is accumulating evidence which identifies the dominant role of genetics in the etiology of both low and high myopia (Francois, p197, 1961; Sorsby, 1962; Goldschmidt, p129, 1968; Duke-Elder, p301, 1970);
2. Environmental elements do not appear to play a role in the development of pathologic myopia (Duke-Elder, p335, 1970). Near work is not considered as an etiologic factor in these cases. The prevalence of pathologic myopia among various occupations requiring a wide range of near work and at various levels of education is basically the same (Goldschmidt, p115, 1968; Curtin, p411, 1985);
3. The severity of posterior fundus changes does not correlate with the degree of refractive error in some instances (Duke-Elder, p300, 314,343, 1970; Duane, V.4, Chap.66, p8, 1984).

Blach et al (1965) indicated that the primary defect of the eye with degenerative myopia is in the layer of the pigment epithelium. Based on their histopathological findings, Blach et al concluded that myopic degeneration is one of the most common tapetoretinal dystrophies. The terms, tapetoretinal degeneration and dystrophy,

are sometimes described in the literature regarding the histopathology of degenerative myopia (Franceschetti, p157,526, 1963). The Latin name of the pigmented epithelium of the retina is "tapetum nigrum" (black carpet, black layer) (Klien, 1950). These terms are deliberately used by a few authors not only to indicate the location and pathologic features of the lesion, but also to suggest its hereditary nature. Several fundus changes are recognized in degenerative myopia. Among the prominent degenerative changes occurring in the maculas of myopic eyes are chorioretinal atrophy, Fuchs' spot and lacquer cracks. Others include pallor and tessellation of the fundus and pigment disturbance. It is not imperative that all of these degenerative changes are present in the same patient or simultaneously, although common features in cases of degenerative myopia can be found, especially if one considers the primary location of these lesions and the anatomy of the macula. Patients with degenerative myopia have a similar clinical course. Marked axial lengthening in these patients often occurs in the early stage of body development, while the posterior fundus changes commence after a lapse of considerable time, especially during young adulthood (Duke-Elder, p301,314,335, 1970; Curtin, p22,37,387,396, 1985; Kanski, p360, 1989). The general ocular picture of these patients is essentially the same although the fundus lesions may vary in size, shape and location, often depending on the stage of development in which the lesions are observed. Degenerative changes in myopic eyes have a tendency to worsen, especially with increasing age (Curtin, p 400, 1985). This seems to suggest that degenerative myopia follows a time-honored development pattern and all of these characteristics may be genetically predetermined. The involvement of the retinal pigment epithelium in degenerative myopia is further confirmed by electrophysiological studies. The EOG value and the amplitude of the ERG b-wave are definitely reduced in most cases of high myopia (Franceschetti, Francois and Babel, p538, 1963; Blach, Jay and Kolb, 1966).

In a clinical study of 120 patients, Jain and Singh (1967) found that the prevalence of abnormal fundi in a unilateral high myopia group was one-half lower than that of a bilateral group. This is

difficult to explain if one assumes that axial elongation and myopic fundus changes are determined by a common gene. The investigators have postulated a genetic imbalance between refractive and degenerative genes in the absence of high myopia in the fellow eye. The point of view that degenerative myopia is a multigenic defect has been supported by a few authors (Francois, 1977, 1961; Duke-Elder, 1970, 1970; Agarwal, Khosla and Angra, 1967).

Earlier studies revealed that retinal pigment epithelium induces the formation of both the choroidal vasculature and the sensory retina (rod and cone cells) during embryological development (Barber, 1955; Duke-Elder, V.3, p73, 1963). The retinal pigment epithelium in human eyes has complex physiological functions, such as photoreceptor cell renewal and Vitamin A transportation, and plays a key role in the normal functioning and health of the retina (Hogan, 1972; Moses and Hart, 1987; Ryan, V.1, p 57, 1989). The task imposed on this single layer of cells is difficult, particularly in the macular region. Any functional breakdown of the retinal pigment epithelium could lead to a variety of degenerative changes in the retina and choroid.

Experimental research has shown that destruction of the retinal pigment epithelium can produce atrophy of the choriocapillaris. Korte and his collaborators (1984), by injecting sodium iodate, destroyed rabbits' retinal pigment epithelium. They found that the choriocapillaris over areas devoid of the pigment epithelium showed severe atrophy while that over areas where the pigment epithelium remained appeared normal. It has been suggested that the choriocapillaris depends on an intact, functioning retinal pigment epithelium for its own survival.

A large-scale study by Curtin and Karlin (1971) showed that the incidence of chorioretinal atrophy increases remarkably with increasing age in patients with same axial length. The older the patient, the more likely s/he will suffer these changes. Chorioretinal degenerations in myopic eyes seem to be inextricably bound to the aging process. It has been confirmed that the residual bodies, originating largely from the outer segment of the photoreceptors, accumulate progressively in the retinal pigment epithelium cells

from early life (Spitznas and Hogan, 1970; Hogan, 1972; Feeney-Burns et al, 1980). Nakaizumi et al (1964) and Hogan et al (1967) demonstrated a marked accumulation of altered collagenous fibers in Bruch's membrane in the elderly. Bruch's membrane becomes weaker and its transmissibility reduces with advancing age. The granular and filamentary materials (residual bodies) are deposited frequently beneath the retinal pigment cells in the majority of older individuals, while forming a continuous layer in others. The individuals belonging to the latter group showed markedly reduced vision when compared with the former group. Sarks (1976), based on a clinicopathological study of 378 eyes, indicated that basal lamina deposits reflect a gradual failure of the retinal pigment epithelium, and is the earliest histopathological manifestation of the age-related degeneration in the macula. In the same study, neovascularization and drusen were found to occur commonly in cases with marked impairment of the pigment epithelium. It is interesting to note that both of these conditions may also be seen in patients with degenerative myopia (Ryan, V.2, p207, 1989; Samuels and Fuchs, p258, 1952). Vogt (1924), according to Duke-Elder (p314,343, 1970), indicated that the resemblance between the histopathological changes characteristic of myopic and senile degenerations is striking. It seems reasonable to postulate that degenerative myopia involves a premature death of a certain cell system. In brief, the heredodegenerative theory considers posterior fundus changes in myopic eyes as an inherited dystrophic process which takes place primarily in the retinal pigment epithelium, and other degenerative changes occurring in Bruch's membrane, choriocapillaris and sensory retina secondary lesions.

PATHOGENESIS

Fuchs' spot is one of the serious complications of degenerative myopia. Unlike other degenerative changes occurring at the posterior pole, the central dark spot can not be directly correlated with increased axial length (Curtin and Karlin, 1971). Forster (1864) first reported this macular lesion found in high myopes. He believed that

it was related to previous retinal hemorrhage. Fuchs (1901) thought that the black spot represented simple pigment accumulation in the macular area. Salzman (1902) demonstrated that many breaks existed in the lamina vitrea of the myopic eye. With the advent of fluorescein angiography, the macular lesion was further linked to subpigment neovascularization. Earlier pathological studies revealed that ingrowth of subpigment new vessels is common in eyes with senile disciform degeneration of the macula (Verhoeff and Grossman, 1937; Maumenee, 1959). Gass (1967) was the first to interpret Fuchs' spot as a disciform response in myopia.

It has been assumed that the disciform response occurring in the macula of the myopic eye involves the following sequence of events :

(1) Predisposed by causative factors, such as metabolic disorders in the pigment epithelial cells, deposition of altered collagenous fibers in Bruch's membrane and mechanical stretching, breaks in the continuity of Bruch's membrane and a loss of normal adherence of RPE to the membrane occur;

(2) New vessels and fibrous tissue invade the subpigment epithelial space from the choroid through defects in Bruch's membrane and form subpigment neovascular membranes (SNM);

(3) Blood or serum may leak continuously from these thin-walled vessels. In many cases, hemorrhagic or serous exudation may further invade the subretinal space, leading to a more extensive retinal detachment in comparison with the primary lesion;

(4) The hemorrhage which forms between the pigment epithelium and Bruch's membrane will eventually undergo organization. Proliferation of the pigment epithelial cells within the mound forms a typical clinical picture of the Forster-Fuchs' spot.

It is not clear at this time what causes subpigment neovascularization in the myopic eye. Defects in Bruch's membrane alone are insufficient to induce the formation of SNMs (Ashton and Sorsby, 1951). A number of current studies showed that SNMs may be present before the development of serous disciform detachment of RPE (Gass, 1973; Teeters and Bird, 1973; Sarks, 1973,1976,1980). Predisposing factors of vasoproliferation are basal lamina deposition of RPE, qualitative changes of collagen and elastic fibers in Bruch's

membrane, and drusen material (Gregor et al, 1977; Sarks, 1973,1980). The role of ischemia of the outer layers of the retina in these cases, however, remains unknown (Archer and Gardiner, 1981). It is unknown whether or not the new vessels stem from the choriocapillaries in such an eye (Henkind, 1978).

PATHOLOGY

Pathological findings in degenerative myopia are somewhat less impressive as similar changes are often found in other conditions. Most of the research in this area were conducted many years ago. The choriocapillaris, Bruch's membrane and the pigment epithelium have been found to be affected most in these cases. However, the exact mechanism of evolution of the condition can not be ascertained given the limited amount of literature available. The basic alterations in degenerative myopia are described as follows :

(1) The choroid: The changes affecting the choroid are essentially atrophic in nature (Duke-Elder, p314). A diffusely thinned choroid is typical in this case. There is a generalized loss of vascular elements and connective tissue framework in this layer. Small diameter vessels appear to be affected first with their walls being thinner than normal. They may obliterate. The stroma is remarkably decreased (Sommers, p314; Curtin, p259). As the degeneration progresses, small vessels may disappear completely over large areas. The larger vessels may also disappear by the end of the degenerative process (Duke-Elder, p314).

(2) Bruch's membrane: This layer is impaired early in the development of myopic degeneration. Particular changes in high myopia are breaks in Bruch's membrane, called lacquer cracks, which appear ophthalmoscopically as whitish-yellow irregular stripes resembling cracks in lacquer. The retina may fuse with the choroid through these ruptures in the absence of the pigment epithelium, producing a picture identical with a late stage inflammatory lesion (Chonan, 1959; Duke-Elder, p328).

Drusen may also be observed in eyes with high myopia besides senile eyes (Samuels and Fuchs, p257; Bird in Krills', 1977; Sarks,

1980). Histologically, drusen manifest as localized depositions of hyaline-like material lying under the basement membrane of RPE (Sarks, 1980). They are PAS-positive, may contain calcium, and differ from Bruch's membrane by their lighter staining. The overlying RPE cells are often abnormal (Yanoff and Fine, p808).

A general disagreement exists regarding the origin of drusen. Drusen have been considered to be abnormal deposits from RPE cells by Spencer (1965) and Horgan (1967), exudative products from the choriocapillaris by Friedman et al (1963) and Gass (1967), and a pathological autolysis of RPE cells by Farkas et al (1971), Burns (1980) and Ishibashi et al (1986). A positive correlation exists between large soft drusen and ingrowth of subpigment new vessels (Gregor et al, 1977; Sarks, 1980).

(3) The retinal pigment epithelium: Involvement of the pigment epithelium is seen early in the course of myopic degeneration. Hyperpigmentation and hypopigmentation are frequently observed (Curtin, p259). The cells in this layer are enlarged and vacuolated (Sommers, p718). Occasionally, a localized ingrowth of fibrovascular tissue from the choroid develops. It appears as a dirty grey patch at the posterior pole. Following a disciform response, proliferation of RPE cells may create a dark spot in the macula of the myopic eye, i.e., Fuchs' spot. In the advanced stage of degeneration, RPE cells may lose their regular pattern or disappear completely and be replaced by connective tissue (Blach in Krill's, p916).

According to Sarks (1976), the basal lamina deposits differ from drusen (colloid bodies). These deposits lie between the basal plasma infoldings and the basement membrane of RPE cells. The author suggests that the deposits are the earliest histopathologic evidence of degeneration of RPE cells in senile eyes. These may be responsible for the formation of subpigment detachment and breaks of Bruch's membrane (Sarks, 1976; Green and Key, 1977). Relative histopathologic studies in myopic eyes, however, are not available at this time.

(4) The retina: Alterations in the sensory retina are thought to be secondary to the degeneration of the pigment epithelium and the choriocapillaris. The receptor cell layer is attenuated in the affected

area. In some instances, the rod and cone cells may be entirely replaced by glial cells (Ryan, V.2, Chap.66, p202). The degenerative changes usually cease in the inner nuclear layer of the retina. This layer has a different blood supply system.

CLINICAL MANIFESTATIONS

CLINICAL FEATURES

Among a number of posterior fundus changes in the myopic eye, Fuchs' spot is of special interest not only due to its unique appearance but also its marked effect upon central vision.

Fuchs (1901) gave a detailed report of the lesion. In his article, the author placed emphasis upon the sharply circumscribed spot in the macula, its dark color, and the progressive changes which occurred with time. He called the macular lesion "the central black spot". Retinal hemorrhages were observed in only 3 of his 11 cases and with myopia from 3 to 22 diopters in patients whose ages were 16 to 71, with an average of 42 years. Most of these patients complained of sudden metamorphopia followed by a rapid decline in vision.

Aiello and Masters (1953) described an unusual fundus lesion. A grayish, spherical, solid mass arising from the choroid and pushing the retina forward in the macular area was found in a patient with myopia of 20 diopters. The lesion was estimated to be two diopters of elevation and about the same diameter as disc. No retinal hemorrhages were found in this case. The lesion was observed for more than one year with no apparent change. The patient's best corrected visual acuity was 20/200 at that time. The authors emphasized that Fuchs' spot could present as a significantly elevated lesion at the posterior pole.

Lloyd (1954) presented a few new cases involving this particular lesion. Besides the clinical observations, the microscopic findings were included in his discussion. He believed that the ophthalmoscopic appearance of Fuchs' spot varies with the age of the lesion and corresponds to a variety of underlying pathological changes occurring in the focal area. The increase in the pigment might explain the black spot: the exudate overlying the spot would lighten the color at the crest and the decrease in pigment around the defect would produce the lightened zone in this area. As the exudate shrinks and the pigmented mass degenerates, the gray or blue-white scar results. The

lesion was believed by the author to be a part of choroidal degeneration in the macula of the myopic eye.

The role of subpigment epithelial fibrovascular tissue in the formation of Fuchs' spot was emphasized by Gass in 1967. The author believed that the circumscribed dark lesion noted in the macula after sudden loss of central vision could be the result of acute hemorrhagic or exudative detachment of the altered pigment epithelium.

Subpigment neovascularization affects 5% to 10% of patients with myopia over 5 diopters (Campos, 1957; Curtin and Karlin, 1971), and 65% to 78% of new cases are associated with subretinal hemorrhage (Fried et al, 1981; Hotchkiss and Fine, 1981). Such a hemorrhage tends to enlarge initially. It is often seen ophthalmoscopically as a "rim" of subretinal blood around the lesion. Organization of subretinal hemorrhage and proliferation of the pigment epithelium later result in a dark spot at the posterior pole. The lesion is usually round or elliptical, and varies in size from a fraction of a disc diameter to measurements well in excess of this. It is often located in the macula, but may be eccentric to it, and even occurs in the nasal fundus (Curtin, p324).

Binocular Fuchs' spots are not uncommon in patients with high myopia. The frequency of the binocular involvement ranges from 18% to 52% in the particular population (Fried et al, 1981). The fellow eyes are frequently affected in a few days or several years. Although the more myopic eye has a significantly higher risk of being affected than the less myopic eye, relatively lower degrees of myopia do not protect the fellow eye completely in the case of anisomyopia (Fried et al, 1981). Bilateral lesions may differ.

With time, these lesions undergo a gradual breakdown in the structure. The well-demarcated, elevated lesions flatten, their margins become indistinct, and depigmentation occurs. By the end of the process, a Fuchs' spot often lies within a large area of chorioretinal degeneration. A certain amount of pigment left in the area may be the only evidence of its previous existence (Curtin, p325).

The color of the lesion has been reported by different authors to be black, grey, emerald green, yellow or reddish (Curtin, p325).

Among all of these colors, grey is the most common. Fuchs' spot can be defined as any dark spot at the posterior pole of the myopic eye.

EVALUATION OF VISUAL FUNCTIONS

Degenerative myopia is an ocular condition characterized by abnormal visual functions, a number of associated complications, and excessive axial length. Careful evaluation of these patients' visual functions is of both diagnostic and prognostic value and of particular worth during the early stages of the condition.

It is well known that there is a tendency towards reduced corrected vision in high myopes. This is true even in younger age groups who do not have obvious chorioretinal degeneration at the posterior pole (Curtin, p277). Visual acuity measurement should be taken routinely on these patients. The testing conditions should be specified so that it is possible for clinicians to make a valid comparison between current and previous findings.

In the case of disciform response, such a patient may notice the visual distortions caused by subpigment transudation in the macular area. Straight lines may look bent or crooked, and faces appear distorted. The Amsler grid is a useful device to detect such changes in very early stages of the condition. Self-monitoring with the Amsler grid is highly recommended to patients at risk of developing disciform degeneration in the macula (Yannuzzi, 1982) because the earlier subpigment neovascularization is detected, the more likely it can be treated successfully.

Visual field evaluation of the highly myopic eye presents a special challenge. Accurate measurement of the visual field is complicated by the high degree of refractive error of the patient. The best method of correction for the test is the contact lens. The central visual field defects found in degenerative myopia are central, paracentral, and arcuate defects as well as enlargement of the blind spot. Central scotomata caused by chorioretinal degenerations may be absolute or relative. These are usually round in shape, and involve 5 to 20 degrees of the field (Curtin, p350). Fuchs' spot often produces a positive scotoma (Aiello and Masters, 1953; Campos, 1957).

Paracentral scotomata caused by small focal areas of myopic atrophy may be seen as grey spots by the patient and, with small fixational movements, the acuity may vary noticeably. Arcuate defects may be encountered in myopia, which are probably ischemic in nature. Glaucoma must be carefully ruled out in these cases (Curtin, p351). Of the blind spot changes, enlargement is the one most frequently seen. This is due to crescent formation. The border of the blind spot often gives an indication of the nature of the process (Franceschetti, 1974). In general, visual field defects correspond roughly to the fundus picture.

Special attention should be paid to functional defects of these eyes because any one of them is an ominous sign. Other signs and defects follow.

DIAGNOSTIC CONSIDERATIONS

The early detection of fine defects at the posterior pole of the myopic eye is substantially more difficult in the presence of a high degree of refractive error, ocular medium opacities, and widespread fundus changes. Great effort must be taken to reveal any subtle changes in the area.

Three complications occur, not uncommonly, in the macular area of patients with degenerative myopia: retinal hemorrhage, lacquer cracks, and the development of a disciform response.

Retinal hemorrhages can be observed in any age group of myopes, more commonly in the young and middle aged. These may occur as isolated events, particularly in the macular area, but are also found in conjunction with fresh lacquer crack formation. The term "retinal" is used here because the source of the hemorrhage has not been fully established. Fluorescein angiography in these eyes usually does not reveal subpigment neovascular membranes in the focal area. The fundus lesions may be round or irregular in shape. When the foveal areas are involved, a variable degree of visual loss and metamorphopsia may occur. Fortunately, this type of hemorrhage can be absorbed over a period of weeks with little or no after effect

(Blach in Krill's, p931; Rabb et al, 1987). These should be distinguished from a disciform response.

Lacquer cracks are seen ophthalmoscopically as irregular, pale lines at the posterior pole of the myopic eye. These are usually associated with a posterior staphyloma. These lines may be independent of one another or arranged in a stellate pattern. Lacquer cracks are generally considered to be breaks in the retinal pigment epithelium-Bruch's membrane- choriocapillaris complex. It is possible that they originate as mechanical tears in the weakened tissues of the myopic eye (Klein and Green, 1975; Shapird and Chandra, 1985). Significant visual dysfunction can rarely be attributed to these lesions, however. Lacquer cracks may be the sources of subpigment neovascularization or hemorrhage. Under such circumstances, the prognosis is much more serious.

Early ingrowth of subpigment new vessels may be clinically undetectable. Later, the neovascular tissue lying between the RPE and Bruch's membrane appears as dirty grey patches of variable size at the posterior pole (Curtin, p323). Subpigment neovascularization tends to occur in the macular area of the myopic eye (Hampton et al, 1983). The onset of a disciform response is often associated with a rapid nonpainful visual loss and metamorphopsia. The fibrous reaction from this type of disciform response is much smaller than that in a classical disciform macular degeneration (Blach in Krill's, p931).

Careful examination of the fundus of the myopic eye is extremely important. Fundus findings establish the diagnosis of degenerative myopia and are the most reliable basis for the prognosis of the disorder.

FLUORESCCEIN ANGIOGRAPHY

BASIC PRINCIPLES AND INTERPRETATION

Fluorescein angiography is of great value in studying the retinal and choroidal circulatory physiology and fundus pathology. The principle of this technique is to visualize and record fluorescence both in intra- and extra- vascular compartments following intravenous injection of fluorescein dye.

Sodium fluorescein is a crystalline substance with a molecular weight of 376.27. The color of its aqueous solution varies from dark red to yellow-green according to its concentration. The dye has the ability to absorb energy from visible light at a short wavelength and emit (fluoresce) at a longer wavelength. The excitation light, the type that is absorbed and changed, is blue (465-490 nm); the resultant fluorescence, or emitted light, is green-yellow (520-530 nm).

Sodium fluorescein is a pharmacologically inert vegetable dye. After injection into the bloodstream, eighty percent of the fluorescein becomes bound to the protein (albumin), and is not available for fluorescence, but 20% remains free in the blood and is available for fluorescence.

White light from the camera flash is passed through a exciting filter which permits only wavelengths in the blue range to strike the retina. This excites the unbound fluorescein within the blood vessels or the fluorescein that has leaked out of the blood vessels, which then emit green-yellow wavelengths which pass back through the pupil to the camera and are recorded on film. A barrier filter is placed just in front of the film that allows the green-yellow light through but keeps out the reflected blue light. This technique permits resolution of fundus details up to five microns and clinically outlines even the smallest retinal capillaries and fine vascular defects seen only by histopathologists in the past (Duane, V.2, Chap.4, p1; Kanski, p350).

A convenient way of analyzing the angiogram is to observe the appearance of the choroidal flush, to follow the sequence of retinal intravascular fluorescence, and finally to note the appearance, if any,

of extravascular hyperfluorescence. The first true fluorescein appears in the choroid approximately 10 to 12 seconds after injection in young patients and 12 to 15 seconds after injection in older patients. A bright glow may be seen when the dye first enters the choroid. This is referred to as the "choroidal flush". Early choroidal fluorescence is usually faint, patchy and segmental because of numerous branchings of the posterior ciliary artery. A watershed zone between the lateral and medial posterior ciliary artery is often seen traversing the optic disc in a vertical direction. Complete filling of the choroid may not be seen until the retinal arterioles are filled. Choroidal fluorescence, however, is not visible in the macula due to the high concentration of pigment in this area. The macular region remains dark throughout the period of angiography.

The dye first appears in the retina through the central retinal artery. The superior and inferior divisions of the central retinal artery are filled very quickly. The patient's pigmentation has no effect on the visibility of the retinal vessels because of lack of physiological blockage-barrier in front of the retinal vessels. The darker the pigment epithelium, the less visible the choroidal fluorescence will be and the greater the contrast of the retinal vascular fluorescence, i.e., vessels are more easily seen.

After the central retinal artery is filled, the fluorescein flows into the retinal arterioles, then the capillaries, the retinal venules, and finally the retinal veins. Because vascular flow is faster in the center of a lumen than on the sides, the fluorescein seems to stick to the sides, creating the laminar pattern of retinal venous flow. The dark central lamina is comprised of nonfluorescent blood that originates from the periphery, and takes longer to fluoresce because of its more distant location. The choroidal and retinal vasculatures are filled maximally about 20 to 25 seconds after injection, and then slowly empty of fluorescein (Ryan, V.2, Chap.57, p22; Archer in Krill's, V.1, p73).

Common causes of abnormal fluorescence are summarized as following (Yannuzzi et al, 1971; Chopdar, p9) :

Hypofluorescence -- It indicates a reduction or absence of normal retinal or choroidal fluorescence at any part of the fundus. There are two possible causes of hypofluorescence:

(1) Blockage of fluorescence: it is sometimes called masking. Normal retinal or choroidal fluorescence is blocked due to increased density of pigment (i.e., choroidal nevus), deposition of abnormal materials (i.e., hard exudates in the neurosensory retina), superficial retinal hemorrhage or ocular medium opacity;

(2) Filling defects: this is caused by an absence of vascular tissue (i.e., myopic degeneration) or by a complete or partial obstruction of the particular vessels (i.e., central or branch retinal vein occlusion). Normal fluorescence is not seen in these cases because it is not present.

Hyperfluorescence -- It indicates an increase in the intensity of fluorescence of any portion of the fundus. Three main factors contribute to the production of hyperfluorescence:

(1) Leakage and staining: Any permeation of dye beyond the physiological barriers is interpreted as leakage of dye. The fluorescein outside the vessels may be loosely bound to certain ocular structures, such as hard drusen and scar tissue, for a short period of time, or enter a space containing exudate within the eye (i.e., serous detachment of RPE). The first phenomenon is designated as staining, and the second being pooling of dye;

(2) Vascular abnormalities: There is increased concentration of dye at the site of tissue pathology. Ingrowth of new blood vessels under the retinal pigment epithelium is a good example. The lesion has greater vascular space than surrounding tissue;

(3) Window defect: It is due to the absence of normal structures which block fluorescence (the retinal pigment epithelium atrophy). The choroidal fluorescence is more visible in the focal area than elsewhere.

A precise diagnosis is not always possible given the angiogram. Previous fundus examination should crystallize the clinician's thinking in analyzing the information derived from the clinical procedures.

ABNORMAL FLUORESCENCE IN DEGENERATIVE MYOPIA

SUBPIGMENT NEOVASCULAR MEMBRANE :

Fluorescein angiography plays an important role in the detection and localization of SNMs in myopia. Previous studies showed that 5% to 10% of patients with myopia over 5 diopters develop choroidal neovascularization (Campos, 1957; Curtin and Karlin, 1971). The new vessels within the membrane fill in an irregular lacy pattern during the early phase of dye transit, fluoresce brightly during peak eye transit (20-30 seconds after injection), and then leak within 1-2 minutes. The fibrous tissue within the membrane then stains with dye and leads to late hyperfluorescence (Levy et al, 1977).

LACQUER CRACK :

The prevalence of lacquer cracks in degenerative myopia has been reported to be 4.3% by Curtin and Karlin (1971) and 9.2% by Rabb et al (1981) respectively. Fluorescein angiography shows pseudofluorescence with faint hyperfluorescence during the late phase of dye transit (window defect) (Rabb et al, 1987; Klein and Green, 1988). Lacquer cracks are often concentrated in the macular area. The width of these lines varies from 50 to 200 μ m (Archer and Logan in Krill's, V.2, p897).

DRUSEN :

Drusen may also be seen in patients with high myopia. These often lead to local atrophy of the overlying retinal pigment epithelium and produce window defects with fluorescein angiography. The lesions appear early in the angiogram as multiple hyperfluorescent spots. Some large drusen may retain fluorescein dye for a long period of time after most of it has emptied. The size of the hyperfluorescent areas, however, remains unchanged with passing time. The phenomenon indicates that fluorescein dye has not leaked but merely adhered to the lesions (staining) (Kanski, p350; Chopdar, p42).

SEROUS DETACHMENT OF RPE :

Serous detachment of the retinal pigment epithelium may be seen in degenerative myopia (Dalkowska, 1975; Curtin, p325). Patients with such lesions may have symptoms of micropsia,

metamorphopsia, or a small central scotoma. The fundus lesion initially creates a small area of masking of choroidal fluorescence on the angiogram. A few seconds later, a small hyperfluorescent spot appears at the center of the previously masked area. Towards the late phase, a large area of leakage of fluorescein dye may form (Chopdar, p43).

ESTIMATION OF VISUAL PROGNOSIS

Before the clinician attempts to predict the final visual outcome for a patient with degenerative myopia, a distinction between Fuchs' spot and subpigment neovascular membrane should be made because the clinical course of the two conditions may be different.

In a retrospective study, Fried et al (1981) found an improvement in vision in 35%, a deterioration in 37%, and stable vision in 28% of their collected cases with degenerative myopia. The follow-up time ranged from three to fifteen years with a median of five years. It was believed that the final visual outcome was mainly related to three factors: the involvement of the fovea, the recurrence of subretinal hemorrhage, and the persistence of neovascular membrane. Subpigment neovascularization was detected by fluorescein angiography in 77% of 55 eyes.

Avila et al (1984) reported that myopic patients with SNMs had a relatively benign visual prognosis. Visual acuity was found stabilized or improved in about 50% of these cases. Most of the eyes in this study showed nonexudative atrophic scars at the posterior pole by the end of a 2 year period of follow-up.

Hotchkiss and Fine (1981), however, demonstrated a worse visual prognosis since visual acuity deteriorated in about 51% of their myopic patients with SNMs. Thirty-eight percent of the potentially treatable eyes, when first seen, became untreatable in their series. Forty-four percent of all cases progressed to legal blindness. Although the presence of neovascular membranes within the foveal avascular zone led to visual loss in most cases, the authors noted that visual outcome was not uniformly poor. Three of the 14 eyes had visual acuities of 20/40 or better after 2 years of follow-up.

A retrospective study by Hampton et al (1983) demonstrated that myopic patients with disciform degeneration had a relatively poor visual prognosis. Forty-three percent of patients in the study lost two or more lines of vision, while 60% were 6/60 or less at the last follow-up. The subfoveal neovascular tissue was detected by means of fluorescein angiography in 58% of the eyes at the first visit; in 23%, it was juxtafoveal; and in 19%, extrafoveal. There appears to exist a direct relationship between the degree of final visual outcome and the distance of the neovascular tissue from the fovea.

Curtin (1977) found that the disorganized Fuchs' spots frequently lay within an enlarging halo of central chorioretinal degeneration. He believed that it might obliterate the retinal area used for eccentric fixation and cause a proportional loss of visual acuity. Vision of 20/200 or less is common in these eyes. The visual prognosis of the disciform response in degenerative myopia is considered poor by the author.

ARGON LASER PHOTOCOAGULATION TREATMENT

Subpigment neovascular membranes may develop in a variety of macular disorders. Marked central vision loss often occurs when these membranes invade into the fovea or cause serous detachment of the pigment epithelium, subretinal hemorrhage and late fibrovascular scarring. When the new vessels have not invaded the fovea, photocoagulation can be used to destroy the membranes in order to preserve central vision.

Two large, randomized, clinical trials (MMSG,1982; MPSG,1986) showed that argon laser photocoagulation of extrafoveal SNMs 200 to 500 um from the center of the foveal avascular zone was clearly beneficial when compared to no treatment. This applies to age-related macular degeneration, ocular histoplasmosis syndrome, and idiopathic choroidal neovascularization. Therapeutic management of subpigment new vessels in degenerative myopia, however, remains a controversial clinical issue.

The concept that laser photocoagulation of subpigment new vessels may be considered when the membrane reduces or threatens to reduce central vision is supported by a number of studies (Hotchkiss and Fine, 1981; Taner and Ilieva, 1985). In the study of Hotchkiss and Fine (1981), five myopic eyes with SNMs extending to the edge of the foveal avascular zone were observed with a average follow-up interval of 25.5 months. The new vessels had advanced to within the avascular zone in all of the cases. Only one of them had a final visual acuity of 20/40 or better. The rest lost two or more lines on the Snellen chart. In contrast to the findings in the untreated group, a better visual outcome was found in the treated group. It was found that initial visual acuity of 20/20 was preserved in three of the five eyes treated with argon laser photocoagulation. The remaining two eyes retained a visual acuity of 20/200. The authors believed that laser photocoagulation was beneficial in cases with SNMs outside the foveal avascular zone or extending to the edge of the perifoveal capillary network.

Avila et al (1984), however, obtained poorer clinical results from laser photocoagulation therapy. Among the 14 eyes, eleven showed

subpigment new vessels in which fluorescein did not leak beyond the border of the membrane (type I), while three showed moderate leakage (type II). An argon green laser (514.5 nm) was used in the study. SNMs were completely closed in all cases, as documented by fluorescein angiography. Immediately after treatment or one year later, nine eyes with type I neovascular membranes had visual acuities the same as or better than the acuities at pretreatment. By the end of the follow-up period (average 30.2 months), however, visual acuity had deteriorated in six of these eyes. In the three eyes with type II neovascular membranes, the final vision was found to be improved or stable. It was also noted that 91% of type I neovascular membranes were found in eyes with severe grades of myopic chorioretinal degeneration, and 80% of type II neovascular membranes were seen in eyes with mild myopic chorioretinal degeneration. These investigators believed that SNMs in eyes with severe degenerative myopia show little or no leakage of fluorescein because the blood flow in the choroidal vessels is noticeably delayed in these cases (Avetisov and Savitskaya, 1977). When laser photocoagulation is performed on these patients, the final visual outcome is usually poor.

Jalkh et al (1987) obtained similar findings. In their series of 19 eyes, visual acuity was improved in only two eyes (11%), stabilized in four eyes (21%), and deteriorated in 13 eyes (68%). It was noted that all except two eyes with type II neovascular membranes showed spontaneous progressive enlargement of the atrophic photocoagulation scar, which worsened visual acuity in 13 eyes (68%). The authors believed that laser photocoagulation should be applied with extreme caution in cases of subpigment new vessels, particularly type I, in degenerative myopia.

Argon blue-green laser photocoagulation is presently contraindicated for SNMs closer than 200 μ m from the center of the foveal avascular zone (Schatz, 1981; Handelman et al, 1981; Fine and Owens, 1983; Kanski, p353). This restricts the number of eyes suitable for the treatment. The newer types of laser, such as diode (810 nm) infrared and Krypton (647 nm) red laser, may eventually

permit more aggressive treatment near to, or actually at, the fovea (Balles and Puliafito, 1990).

CONCLUSION

Fuchs' spot is one of the major complications of degenerative myopia. Significant visual loss often occurs in these cases. Careful evaluation of these patients' visual functions and fundi is of both diagnostic and prognostic value, and of particular worth during the early stages of the condition. Fluorescein angiography plays a key role in the detection and localization of subpigment neovascular membranes (SNMs). Laser photocoagulation may be beneficial in patients with SNMs outside the foveal avascular zone. It has been known that Fuchs' spot results from two different stages in the development of a hemorrhagic disciform macular detachment in myopia. A complete understanding of the exact mechanism of chorioretinal degenerations in the myopic eye must await future research.

APPENDIX

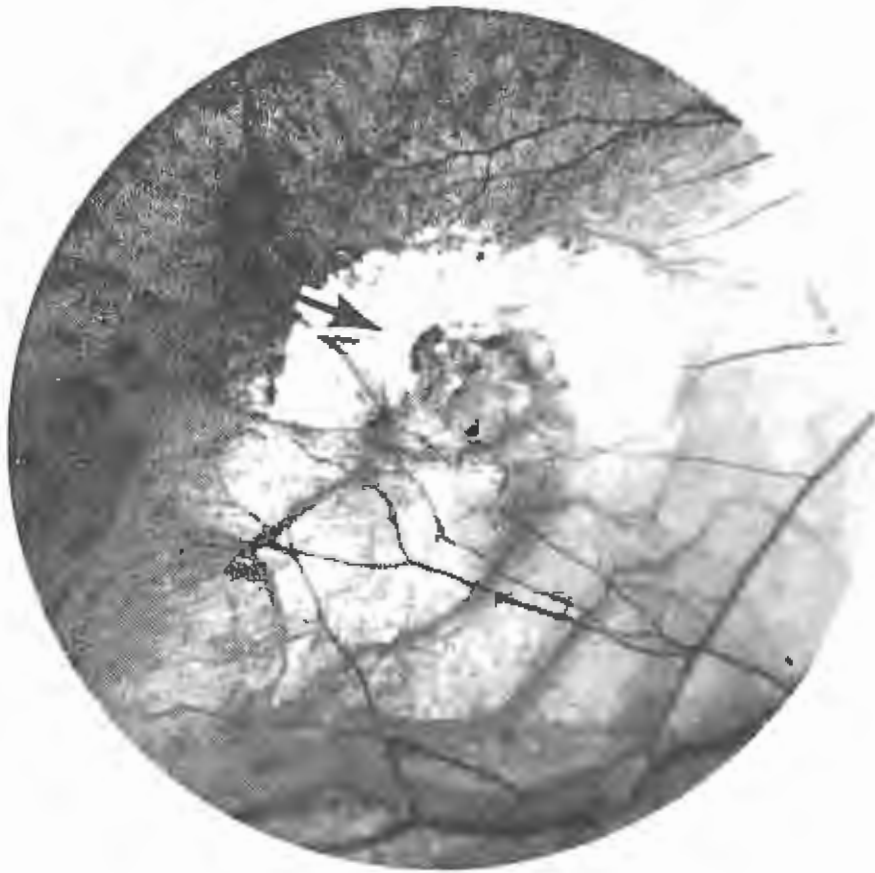


FIG. 1

Small pigmented, neovascularized, subretinal scar (Fuchs' spot) in a patient with high myopia (arrow).

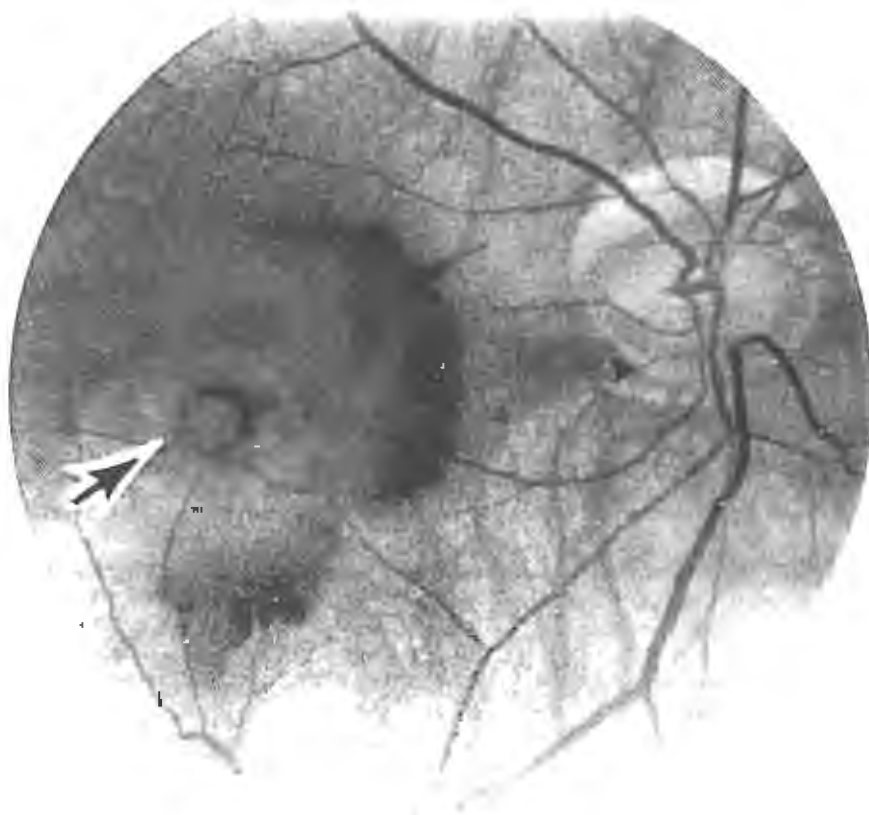


FIG. 2

Incompletely resolved hemorrhagic detachment of the macula surrounding a pigmented mound in an area of choroidal neovascularization (arrow).

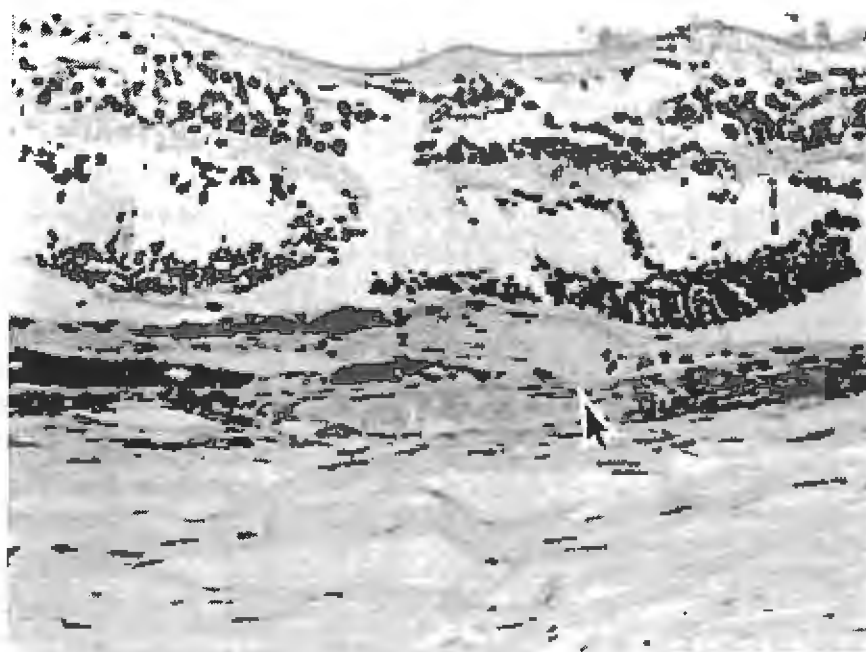


FIG. 3

Histopathology of Fuchs' spot in degenerative myopia. Note thin choroid and submacular fibrovascular scar (arrow).

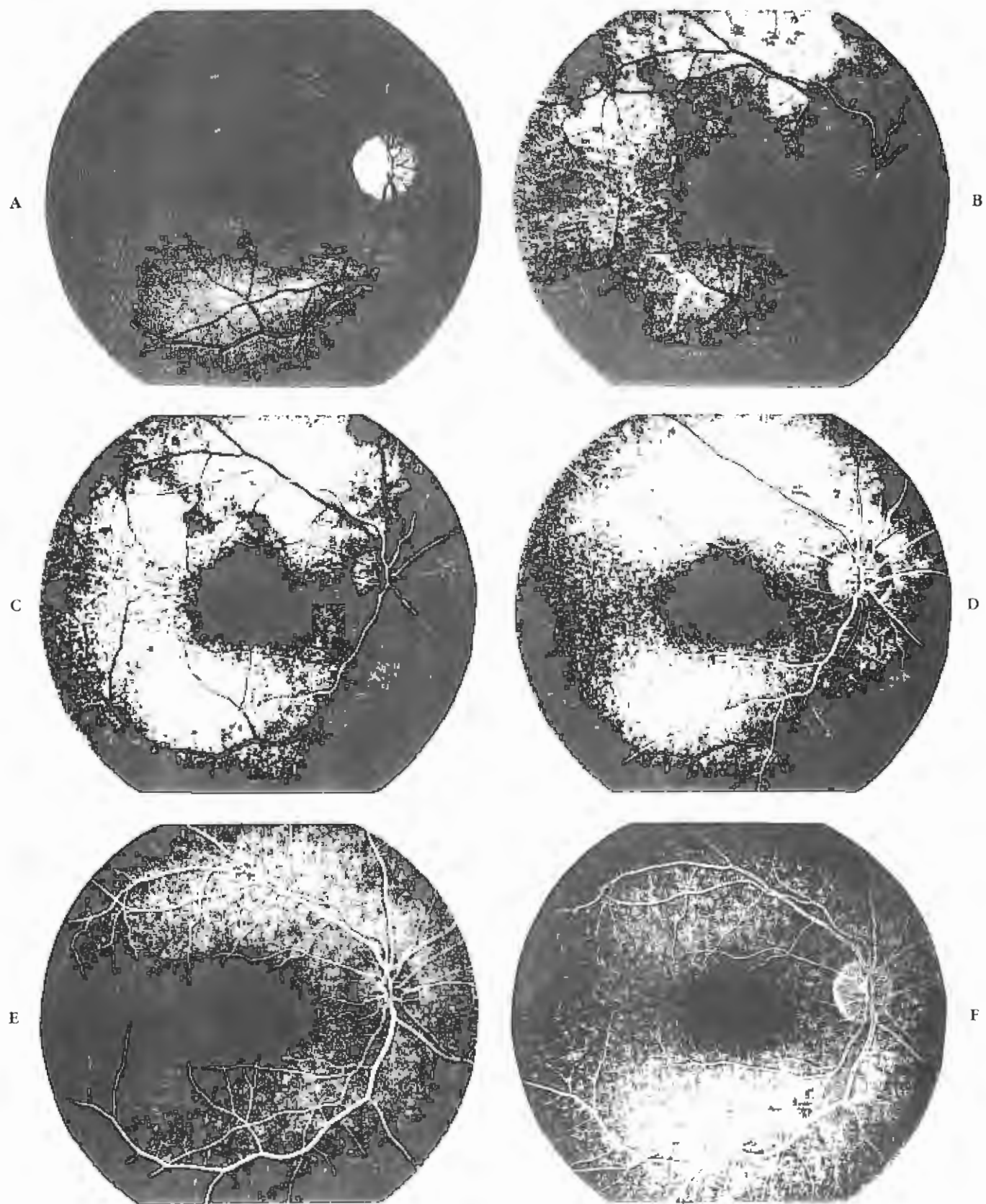


FIG. 4

FIG. 4 Normal fluorescein angiogram of right disc and macula taken with a 60-degree camera. **A**, Red-free photograph. The disc and macula are normal. The reflex above and below the foveal area is normally seen in a young patient. **B**, The ground-glass fluorescence is the very early fluorescein filling of the choroid—the *choroidal flush*, which often occurs a few seconds before fluorescence within the retinal arteries, which in this photograph are just beginning to fill with fluorescein. **C**, Early arterial phase of angiogram. Note that the retinal arteries are filling, and the retinal veins have not yet begun to fluoresce. The choroid is almost completely fluorescent. The dark patches are areas of hypofluorescent choroid that have not yet received fluorescein; this is called *patchy choroidal filling*. **D**, Early arteriovenous phase of the angiogram. The retinal veins have begun to show fluorescein filling, as evidenced by the laminar flow within the veins. The patchy choroidal filling has mostly cleared, although still remaining are two patches faintly evident above the disc, one large patch below, and one nasally. **E**, Mid-arteriovenous phase of the angiogram. The choroid has completely filled with fluorescein, as have the retinal arteries and veins. The macula remains dark. **F**, Late phase of the fluorescein angiogram shows that fluorescein has faded from both the choroid and retinal vessels. There is very slight staining along the disc margin inferotemporally. The macula has remained dark.

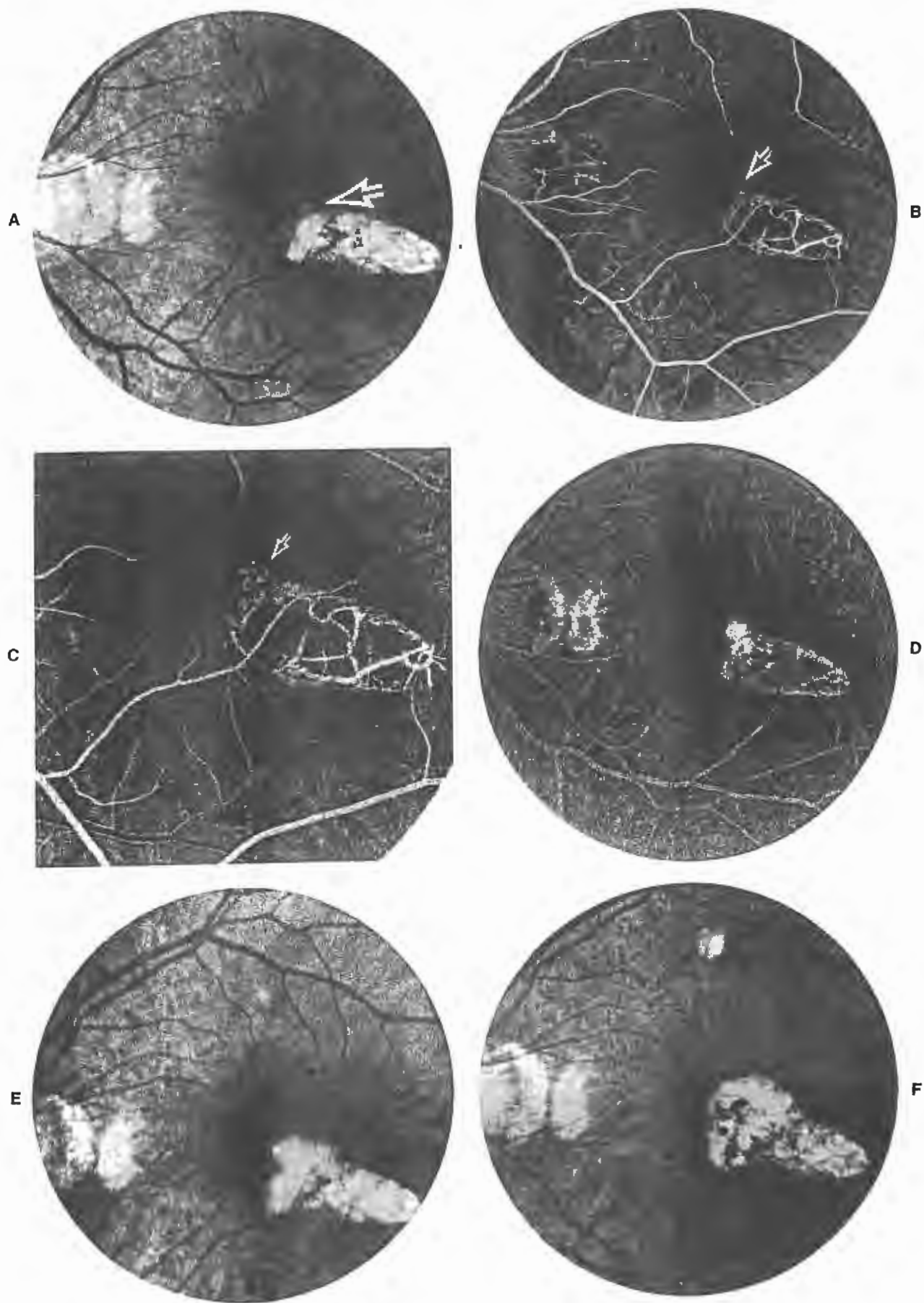


FIG. 5

FIG. 5

A, Serous detachment of the macula secondary to choroidal neovascularization occurring at the margins of an area of chorioretinal atrophy in a 30-year-old man with high myopia. Note the pigment ring (arrow) at the superior margin of the atrophic choroidal lesion. **B** and **C** (magnified view), Early angiogram shows evidence of a small choroidal neovascular membrane (arrow) just on the temporal edge of the capillary-free zone of the foveolar region. **D**, Late angiogram shows staining in the region of the choroidal neovascularization. **E**, The choroidal neovascular membrane and its feeding vessels were treated with argon laser photocoagulation. **F**, Two years following photocoagulation treatment. The patient's visual acuity was 20/20.

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